Auris Nasus Larynx xxx (2016) xxx–xxx

Contents lists available at ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

Free-flap reconstruction for the management of life-threatening hereditary hemorrhagic telangiectasia

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ARTICLE INFO

Article history:
Received 8 April 2016
Accepted 13 August 2016
Available online xxx

Keywords:
Hereditary hemorrhagic telangiectasia
Osler–Rendu–Weber syndrome
Epistaxis
Free flap
Septodermoplasty

ABSTRACT

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant multi-systemic disease that exhibits increasing penetrance with age. Some patients present with severe life-threatening epistaxis which is intractable to all common treatment modalities.

A 63-year-old female patient with hereditary hemorrhagic telangiectasia (HHT) presented with recurrent life-threatening epistaxis that required repeated transfusions despite multiple embolizations and septodermoplasty. Previous septodermoplasty failed due to septal perforation. Resurfacing of the nasal lining with a free flap was planned. Total removal of the nasal mucosa and remaining septum was conducted to make the nasal cavity into one common cavity. Nasal passages were resurfaced with a radial forearm free flap. Following surgery, the patient experienced no further significant epistaxis.

Fasciocutaneous free-flap resurfacing might represent a curative solution for cases of HHT intractable to conservative treatment and septodermoplasty, especially for patients with large septal perforation.

1. Introduction

Hereditary hemorrhagic telangiectasia (HHT), also known as Rendu–Osler–Weber syndrome, is an autosomal-dominant, multi-systemic disease that shows increasing penetrance with age. Its clinical symptoms include epistaxis, mucocutaneous and gastrointestinal telangiectasia, and internal arteriovenous malformations in the lung, brain, or liver [1]. Its prevalence is between 1:5000 and 1:8000. Different types of mutations in various loci are thought to cause HHT [2,3].

The most frequent clinical manifestation of HHT is epistaxis (nose bleeding), which affects 93% of patients and is normally light to moderate. The management of HHT typically includes general measures, iron supplementation, occlusive treatments, surgery, interventional radiology, and/or pharmacological treatments [4,5]. Septodermoplasty is a surgical option for severe HHT patients and is very effective when the main bleeding is at the nasal septal mucosa or floor of the nasal cavity. However, when the main bleeding is in the lateral nasal wall, or when the patient has septal perforation, resurfacing the problematic mucosa with skin is technically difficult. In these cases, effective covering of the convoluted lateral wall and midline mucosa is challenging due to the presence of turbinate and septal perforation. Here, we report a patient with life-threatening severe epistaxis that was intractable to medical treatment and who experienced septodermoplasty failure. This patient was successfully treated by resurfacing of the nasal mucosa with forearm free-flap reconstruction using microsurgical techniques.

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http://dx.doi.org/10.1016/j.anl.2016.08.005
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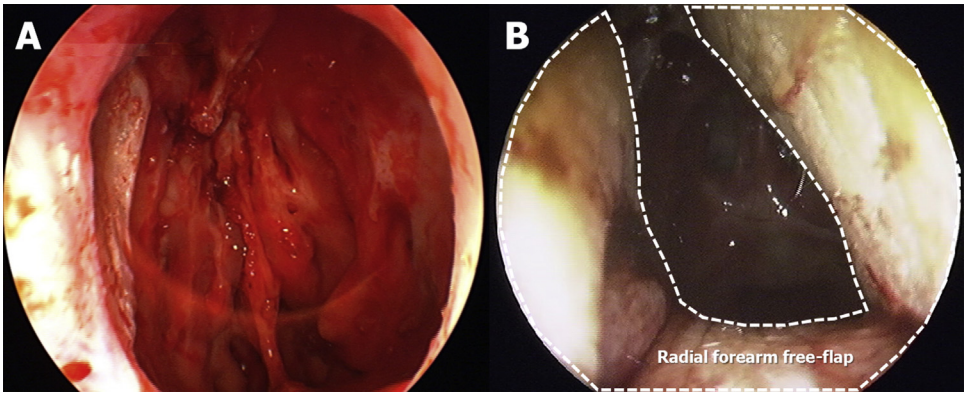

Fig. 1. Pre- and postoperative view. (A) One common nasal cavity with a large septal perforation was observed preoperatively. (B) The lateral nasal wall resurfaced with a tubular shaped radial forearm free-flap (dotted line) was observed 1 year later after operation.

2. Case report

A 63-year-old female patient presented with recurrent life-threatening epistaxis that required repeated transfusions. Hemoglobin levels ranged from 4.2 to 7.4 grams per deciliter after each event. In addition, she showed telangiectasia in distal parts of her fingers, toes, tongue, and lower lip with frequent bleeding. She had a confirmed mutation in *ENG*, which encodes endoglin, and two of her three children had been diagnosed with HHT caused by the same mutation. She had undergone surgery for pulmonary arteriovenous malformation 12 year ago. The severe recurrent epistaxis events began 10 years earlier and the bleedings were controlled with conservative management such as nasal packing and cauterizations. She was referred to our department in 2013 for surgical correction. In her initial endoscopic examination, she presented with a large septal perforation caused by frequent cauterization and excessive packing performed at other clinics (Fig. 1A). We planned septodermoplasty to replace the mucosa of the lateral wall, nasal floor, perforation margin, and remaining septal mucosa into the skin, although we expected difficulty due to loss of midline septal support caused by septal perforation. Septodermoplasty for patient with septal perforation was performed according to the previously described technique [6]. Using scissors and cutting forceps, the residual nasal septum was resected for making U-shaped one nasal cavity. The mucosa of the lateral nasal wall and nasal floor was removed. The septodermoplasty was then conducted with split-thickness skin graft from the right thigh. One week later after the surgery, massive bleeding occurred after packing removal, and we determined that the skin covering the nasal floor, septal mucosa, and lateral nasal wall should be removed again to control bleeding. The operation for bleeding control was performed under general anesthesia, grafted skin was partially preserved at the lateral nasal wall, caudal to the inferior and middle turbinate and further resection of the nasal septum and middle turbinate was performed, with a concept of removing the end organ. Despite the surgical resection, several times of electrocauterization was repeated for bleeding control. Two months later, we planned arterial embolization to control repeated epistaxis. She underwent transfemoral external carotid arteriography and hypervascular stainings were identified in distal part of both internal maxillary artery and extraorbital branch of ophthalmic artery. Both distal IMA branches and right ophthalmic artery were embolized using polyvinyl chloride and gelfoam particle. She was also treated with iron supplements, tranexamic acid, and tamoxifen, which are known to be effective treatments to reduce bleeding.

Despite these efforts, she had continued epistaxis and required repeated transfusions for 2 years. The surgical team decided on the more drastic measure of lining the nasal mucosa with a free flap. The entire nasal mucosa was stripped out, and

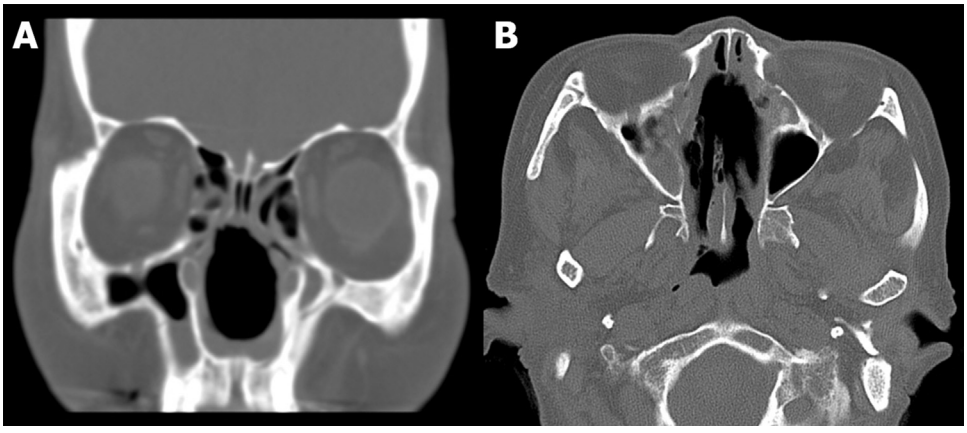

Fig. 2. Preoperative computed tomography. One common nasal cavity with a large septal perforation was observed in the coronal (A) and axial view (B).

Please cite this article in press as: Choi JW, et al. Free-flap reconstruction for the management of life-threatening hereditary hemorrhagic telangiectasia. Auris Nasus Larynx (2016), http://dx.doi.org/10.1016/j.anl.2016.08.005